General

Guideline Title

Care of the patient with myasthenia gravis.

Bibliographic Source(s)

American Association of Neuroscience Nurses (AANN). Care of the patient with myasthenia gravis. Glenview (IL): American Association of Neuroscience Nurses (AANN); 2013. 33 p. [108 references]

Guideline Status

This is the current release of the guideline.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- August 31, 2016 – Opioid pain and cough medicines combined with benzodiazepines : A U.S. Food and Drug Administration (FDA) review has found that the growing combined used of opioid medicines with benzodiazepines or other drugs that depress the central nervous system (CNS) has resulted in serious side effects, including slowed or difficult breathing and deaths. FDA is adding Boxed Warnings to the drug labeling of prescription opioid pain and prescription opioid cough medicines and benzodiazepines.
- March 22, 2016 – Opioid pain medicines : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

Recommendations

Major Recommendations

The levels of recommendation (1-3) and the data quality classifications (I-IV) are defined at the end of the "Major Recommendations" field.

Ocular Only Versus Generalized Myasthenia Gravis (MG)

Ocular MG

Nurses should monitor the results of the ice pack test with consideration for its limited value in the diagnosis of MG (Level 2).

Diagnostic Tests

Laboratory Tests

Nurses should know the adverse events associated with diagnostic testing in MG and how to manage those side effects as needed. Nurses should monitor for results and adverse events associated with the use of edrophonium for diagnostic testing in MG and manage adverse events appropriately (Level 2).

Electrodiagnostic Testing

Nurses should be familiar with electrodiagnostic testing in MG. Nurses should prepare patients for their experience, including the likelihood of some discomfort (Level 2).

Other

Nurses should understand and explain the "ice pack test" and diagnostic imaging to the patient and family to lessen fear and anxiety and elicit cooperation and participation (Level 2).

Nursing Assessment of the Patient with MG

Assessment Techniques

Nurses should perform a comprehensive nursing assessment of the patient with MG that includes a detailed medical history and symptom description. Nurses should test all voluntary muscles for both muscle strength and fatigability to determine the severity of specific muscle weakness, the degree of functional impairment, and potential complications (see Table 4 in original guideline document) (Level 3).

Respiratory Muscles

Nurses should perform a detailed physical assessment of respiratory status (Level 3).

Nurses should assess the respiratory system appropriately and support respirations as needed until additional interventions are available (Level 3).

Treatment

Pharmacological Management

Nurses must administer and teach the patient with MG and their family to administer the correct dose of acetylcholinesterase (AChE) inhibitors on time (Level 2). Nurses must also teach the patient and their family the signs and symptoms of overdose and adverse events (Level 2).

Nurses should advise patients who are taking corticosteroids to be aware of infection risk, practice good hand hygiene practices, and avoid exposure to people with infection (Level 2).

Nurses should monitor for side effects associated with immunosuppressants and minimize patient risk for infection by maintaining appropriate infection-control practices, including good hand hygiene and sterile technique for invasive procedures (Level 2). Nurses should administer antimicrobials as prescribed (Level 2).

Nurses should monitor for signs and symptoms of adverse effects associated with all medications used in the management of MG as previously described, including those most commonly associated with long-term immunosuppressants/ immunomodulating agents, specifically, infection, hepatotoxicity, nephrotoxicity, bone marrow suppression, and skin cancer (Level 3).

Other Therapies

Plasmapheresis (Therapeutic Plasma Exchange [TPE])

Nurses should monitor for improvement following plasmapheresis (Level 2).

Nurses should monitor for complications associated with venous access (Level 2).

Nurses should monitor for side effects associated with plasmapheresis including fatigue and hypotension (Level 2).

Intravenous Immunoglobulin G (IVIG) Infusions

Nurses must understand the rationale for the use of IVIG in the management of MG and its reported benefits and limitations (Level 2). Nurses must be knowledgeable of different brands of IVIG and institution-specific protocols regarding dosage and rates, which may include a test dose and then a gradual increase in rate as tolerated to lessen risk for adverse effects. Action plans for adverse effects must be followed (Level 2).

Thymectomy

In collaboration with the multidisciplinary team, nurses should educate the patient and family regarding indications, pre- and postoperative care, and prognosis for patients undergoing thymectomy (Level 3). Following thymectomy, nurses should monitor the patient closely for complications related to MG and the thymectomy procedure and manage them effectively (Level 3).

Postoperative Care After Thymectomy

Nurses should monitor respiratory status, including rate, depth, work of breathing, breath sounds, oxygenation, and ventilation tolerance. Spontaneous breathing trials, pulmonary function studies, forced vital capacity (FVC), and negative inspiratory force (NIF) should be ordered as available (Level 3).

Nurses must administer MG medications postoperatively as prescribed. Nurses should balance the administration of medications that may worsen MG, including opioids, with a comfort level that will allow the patient to breathe with the least amount of effort and discomfort (Level 3).

Nurses should follow the manufacturer's direction for the chest tube drainage system and monitor chest tube(s) for patency, underwater seal, evidence of air leak, drainage output, and signs and symptoms of infection. Nurses should monitor for signs and symptoms of increasing pneumothorax or hemothorax (Level 3).

Nurses should be aware of patients who have factors that may increase the likelihood of a postoperative MG crisis that could complicate their recovery (Level 3).

Nurses should monitor closely for signs and symptoms of infection and minimize a patient's risk for infection by consistently complying with infection-control measures (Level 3).

MG Crisis

Nurses should be knowledgeable in the differentiation of the two different types of MG crises, myasthenic crisis or cholinergic crisis, and should recognize that both are care priorities. In either situation, nurses should perform a complete respiratory and neuromuscular assessment, which is essential to identify ineffective respiratory function and impaired gag and swallow, and initiate the appropriate airway-management strategies and oxygen delivery (Level 3) (Vassar et al., 2008).

Nurses should assess respiratory status, including pulmonary function tests (i.e., NIF and FVC); provide pulmonary hygiene as needed; and alert the licensed independent provider regarding indications for additional respiratory therapy management to minimize respiratory complications, including prolonged ventilation and pneumonia (Level 3).

General Nursing Management of the Patient With MG

Nurses should administer drugs that may worsen MG with caution. Review medication profiles with the pharmacist and licensed independent provider (Level 3).

Nurses should adopt the previous protocols to facilitate swallowing, avoid aspirations, and optimize nutritional and fluid status (Level 3).

Nurses should apply the above strategies in managing fatigue for the patient with MG (Level 3).

Special Consideration in the Juvenile MG Population

Nurses should be aware of the teratogenic effects of immunosuppressive medications on the fetus and provide appropriate education to women with MG of childbearing age (Level 3).

Psychosocial and Educational Needs

Nurses should educate the patient with MG and their family regarding management of the disease; influence of the disease on lifestyle, swallowing, and chewing impairment; and fatigue and energy conservation (Level 3).

Definitions:

Data Quality Classification

Class I: Randomized controlled trial (RCT) without significant limitations or meta-analysis

Class II: RCT with important limitations (e.g., methodological flaws or inconsistent results) and observational studies (e.g., cohort or case-control)

Class III: Qualitative study, case study, or series

Class IV: Evidence from expert committee reports and/or expert opinion of the guideline panel, standards of care, and clinical protocols

Levels of Recommendation

Level 1: Recommendations are supported by class I evidence.

Level 2: Recommendations are supported by class II evidence.

Level 3: Recommendations are supported by class III and IV evidence.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Myasthenia gravis (MG)

Guideline Category

Counseling

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Critical Care

Family Practice

Internal Medicine

Neurology

Nursing

Thoracic Surgery

Intended Users

Advanced Practice Nurses

Hospitals

Nurses

Guideline Objective(s)

- To review and evaluate literature about myasthenia gravis (MG), with a concentrated focus on the adult with acquired MG, and to create a reference for neuroscience nurses who care for the patient with MG across the continuum of care throughout the lifespan
- To offer evidence-based recommendations on nursing activities that have the potential to maximize outcomes for patients of all ages and all types of MG
- To help nurses provide consistent, current, and evidence-based care to the patient with MG

Target Population

Adults with acquired myasthenia gravis (MG)

Interventions and Practices Considered

Diagnosis/Evaluation

1. Understanding and preparing patients for diagnostic tests, including ice pack test, laboratory tests, and electrodiagnostic tests
2. Nursing assessment of the patient with myasthenia gravis (MG)
    - Medical history and symptom description
    - Testing of all voluntary muscles for both muscle strength and fatigability
    - Detailed physical assessment of respiratory status and respiratory system

Treatment/Management

1. Pharmacological management, including administrating, monitoring, and teaching patients about acetylcholinesterase (AChE) inhibitors, corticosteroids, and immunosuppressants/immunomodulating agents
2. Monitoring for improvement, complications, and side effects following plasmapheresis
3. Understanding the rationale for the use of intravenous immunoglobulin G (IVIG) and becoming knowledgeable of different brands of IVIG and institution-specific protocols regarding dosage and rates
4. Educating the patient and family regarding indications, pre- and postoperative care, and prognosis for patients undergoing thymectomy
5. Monitoring and managing the patient closely for postoperative complications
6. Understanding MG crisis and initiating appropriate airway management strategies
7. Employing general management strategies, including adopting protocols to facilitate swallowing and strategies to manage fatigue

Major Outcomes Considered

- Sensitivity and specificity of diagnostic tests
- Exacerbation and remission of myasthenia gravis (MG)
- Value of history and assessment techniques
- Effectiveness of medications and other treatments for symptoms and disease management
- Adverse effects of treatment
- Surgical and postoperative outcomes (thymectomy)
- Benefits of psychosocial and educational counseling

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

A review of literature published between January 2000 and September 2012 was conducted using PubMed/MEDLINE and CINAHL databases with the following search terms: myasthenia gravis, neuromuscular junction (NMJ), neuromuscular transmission, thymectomy, evidence-based practice nursing, research, and myasthenia gravis clinical trials. The search was extended to earlier years because of the lack of more recent references on select topics. Several publications dated earlier than 2000 are included because of their historical clinical significance.

The Myasthenia Gravis Foundation of America guidelines were assessed and incorporated in this document as appropriate and needed.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Data Quality Classification

Class I: Randomized controlled trial (RCT) without significant limitations or meta-analysis

Class II: RCT with important limitations (e.g., methodological flaws or inconsistent results) and observational studies (e.g., cohort or case-control)

Class III: Qualitative study, case study, or series

Class IV: Evidence from expert committee reports and/or expert opinion of the guideline panel, standards of care and clinical protocols

Methods Used to Analyze the Evidence

Systematic Review

Description of the Methods Used to Analyze the Evidence

Not stated

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

The *Clinical Practice Guidelines* and recommendations for practice are established based upon the evaluation of the available evidence. Resources and recommendations must describe the best practices that can enable registered nurses (RNs) to provide optimal care for persons with myasthenia gravis (MG).

Rating Scheme for the Strength of the Recommendations

Levels of Recommendation

Level 1: Recommendations are supported by class I evidence.

Level 2: Recommendations are supported by class II evidence.

Level 3: Recommendations are supported by class III and IV evidence.

Cost Analysis

A cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Peer Review

Description of Method of Guideline Validation

The guideline underwent peer review by a panel of reviewers listed in the guideline document.

Evidence Supporting the Recommendations

References Supporting the Recommendations

Vassar TM, Batenjany M, Koopman WJ, Ricci M. Nursing issues. In: Howard JF, editor(s). Myasthenia gravis: A manual for the health care provider. 1st ed. St. Paul (MN): Myasthenia Gravis Foundation of America; 2008. p. 32-53.

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

The recommendations may assist nurses in making appropriate choices when caring for patients with myasthenia gravis (MG).

Potential Harms

- Adverse events associated with certain diagnostic tests, including the use of edrophonium for diagnostic testing
- Adverse effects or events associated with pharmacological management (e.g., fasciculations, abdominal cramps, and diarrhea with acetylcholinesterase [AChE] inhibitors; myelosuppression, infection, hepatotoxicity, nephrotoxicity, and skin cancer with immunosuppressive/immunomodulating drugs)
- Adverse effects associated with plasmapheresis including fluid volume changes and hypocalcemia from citrate used during plasmapheresis, fluid overload, congestive heart failure, infections, and thrombotic and bleeding tendencies
- Adverse reactions associated with intravenous immunoglobulin G (IVIG) infusions, including circulatory overload, renal failure, nausea and

vomiting, headaches, aseptic meningitis, thrombosis, stroke, seizures, retinal vasculitis, skin rashes, and alopecia
- Adverse events associated with crisis or surgical care or treatment, such as ventilator care

Contraindications

Contraindications

- It may be necessary to avoid azathioprine, cyclosporine, mycophenolate, and cyclophosphamide in women of reproductive age because of possible teratogenic effects on the fetus.
- Intravenous edrophonium chloride is contraindicated in the patient with bronchial asthma or cardiac dysrhythmias because edrophonium may result in cholinergic side effects.

Qualifying Statements

Qualifying Statements

- The authors, editors, and publisher of this document neither represent nor guarantee that the practices described herein will, if followed, ensure safe and effective patient care. The authors, editors, and publisher further assume no liability or responsibility in connection with any information or Recommendations contained in this document. These Recommendations reflect the judgment from the American Association of Neuroscience Nurses regarding the state of general knowledge and practice in the field as of the date of publication and are subject to change based on the availability of new scientific information.
- Adherence to these guidelines is voluntary, and the ultimate determination regarding their application must be made by practitioners in light of each patient's individual circumstances. This reference is an essential resource for nurses providing care to the patient with myasthenia gravis (MG). It is not intended to replace formal learning but rather to augment the clinician's knowledge base and provide a readily accessible reference tool.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

American Association of Neuroscience Nurses (AANN). Care of the patient with myasthenia gravis. Glenview (IL): American Association of Neuroscience Nurses (AANN); 2013. 33 p. [108 references]

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2013

Guideline Developer(s)

American Association of Neuroscience Nurses - Professional Association

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Guideline Committee

Not stated

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Financial Disclosures/Conflicts of Interest

A staff member, volunteer or family member who has an affiliation with a commercial company must declare that relationship in advance of involvement with an education activity that could be used as a forum to promote interest of that company and its products. None of the authors or planners has anything to disclose related to this activity.

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the American Association of Neuroscience Nurses Web site .

Availability of Companion Documents

None available

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on August 29, 2013. The information was verified by the guideline developer on September 14, 2013. This summary was updated by ECRI Institute on June 2, 2016 following the U.S. Food and Drug Administration advisory on Opioid pain medicines. This summary was updated by ECRI Institute on October 21, 2016 following the U.S. Food and Drug Administration advisory on opioid pain and cough medicines combined with benzodiazepines.

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